OP0079

LIMB GIRDLE MUSCULAR DYSTROPHY TYPE 2B - A RARE MYOSITIS MIMIC

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Background: Proximal muscle weakness with associated raised creatine kinase (CK) commonly leads to referral to Rheumatology for the investigation of Idiopathic Inflammatory Myopathy (IIM). Some genetic myopathies can have a similar presentation with investigations that suggest inflammatory disease, leading to difficulty with accurate diagnosis (Amato & Brown, 2011; Harlan & Mammen, 2019).

Objectives: To describe the case of a patient with Limb Girdle Muscular Dystrophy Type 2B (LGMD2B), whose initial presentation mimicked an inflammatory myopathy.

Methods: Case report.

Results: A 43-year-old patient was reviewed by Rheumatology due to proximal muscle weakness with a raised CK. Muscle biopsy was suggestive of inflammatory myopathy. Therefore, he was started on treatment with corticosteroids. Corticosteroid treatment resulted in no improvement in his weakness or CK. His diagnosis was reviewed, and he was referred to the Neurology and Genetics services. Following molecular genetic analysis, a diagnosis of Limb Girdle Muscular Dystrophy Type 2B was made.

Conclusion: Muscle biopsies can suggest an inflammatory aetiology in some genetic myopathies (Amato & Brown, 2011; Harlan & Mammen, 2019). If a patient with suspected IIM presents with atypical features, or they do not respond as expected to treatment, then consider a genetic myopathy such as LGMD2B as a cause and involve the Neurology and Genetics services in the case.

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OP0080

OSTEOPOIKILOSIS IN PATIENT WITH ANKYLOSING SPONDYLITIS

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Background: Osteopoikilosis (OPK) – a rare congenital genetic-mediated benign sclerosing disease of skeleton. OPK is usually clinically asymptomatic and is most often an incidental finding during an X-ray study [1]. The lesions are from 2 to 10mm and are located in the area of the ischial, pubic bones, metaphyseal regions of the tubular bones. The axial skeleton is usually not affected in OPK. As early described, OPK may be associated with inflammatory rheumatic diseases, such as rheumatoid arthritis, spondyloarthritis, reactive arthritis and scleroderma [2–4]. Ankylosing spondylitis is a chronic rheumatic disease from the group of spondyloarthritis with obligatory involvement of the sacroiliac joints and frequent involvement of entheses and peripheral joints. Although a significant mutual influence of OPK and AS has not been proven, their combination can lead to unreasonable over-examination in real clinical practice.

Objectives: To present a clinical case of rare coexisting of osteopoikilosis and ankylosing spondylitis.

Methods: Case report. The patient was treated and examined based on real clinical practice.

Results: A 33-year-old man was admitted to our rheumatology department with a three-year history of inflammation pain in cervical region of spine and hip, knees and shoulder joints and long morning stiffness duration (up to 3 hours). AS was earlier verified based on modified New York criteria (1984) and Nonsteroidal anti-inflammatory drugs were prescribed promptly. On the outpatient basis he also received sulfasalazine and methotrexate due to arthritis and enthesitis. Nevertheless he had high disease activity (ESR 66mm/h, CRP 77.5mg/l, BASDAI = 7.5), severely limited range of motion in the cervical, thoracic and lumbar spine, arthritis, enthesitis when he entered in our clinic. He had been treated with secukinumab 150mg for fourteen months and then he was switched to golimumab 50mg per month because of lack of efficacy. Low disease activity has now been achieved.

During the assessment of radiographs of the pelvic bones and spine, typical changes of the late stage of ankylosing spondylitis (syndesmophytes, stage 4 sacroiliitis) were revealed at the first visit in our clinic. At the same time, a number of phenomena were discovered that were not related to the course of the AS, which required expanding the differential diagnosis range. The tumor process and endocrine pathologies were excluded. OPK was diagnosed on the basis of characteristic changes in various parts of the skeleton, especially in phalanges, wrists and pelvis bones (fig.1).

Figure 1. OPK spots in phalanges and wrists.

Conclusion: We've reported rare case of AS and OPK coexisting in young patient. The identification of typical radiological phenomena and correct evaluation of clinical data may allow to avoid unnecessary expensive examinations.

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OP0081

A CASE OF ATYPICAL MYCOBACTERIUM INFECTION COMPLICATING EXTRA-NODAL ROSAI-DORFMAN DISEASE IN A PATIENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS (SLE)

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Background: We present the case of a 28 year old Black-British female with severe SLE requiring treatment with rituximab in 2012 due to persistent low-grade activity and severe episodes of pleuro-pericardial effusions, pancytopenia, fever and weight loss